

## Ultrasonographic Display of Complex Vascular Rings

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Ultrasonographic display of vascular ring anatomy has been limited to single-plane views. This does not readily allow for a three-dimensional interpretation of structural relations. A method that included a sweep consisting of multiple contiguous frontal planes was used in 12 patients with a vascular ring before repair for the evaluation of the arch sidedness and number, brachiocephalic vessel pattern, upper descending thoracic aorta sidedness, ductus arteriosus site or sites and proximal pulmonary arteries; 7 patients had Doppler color flow imaging.

Complete imaging of the luminal vascular components

was possible in all but one patient. In four other patients, atretic segments of the vascular ring could not be displayed. The addition of Doppler color flow imaging especially aided in the tracing of multiple vascular structures in complex cases and in assessing ductus arteriosus and arch patency. The use of a suprasternal frontal sweep with posterior angulation could display encirclement of the air-filled trachea. Vascular ring segments without lumens could not be displayed.

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Reports (1-10) of ultrasonographic methods to assess the anatomy of vascular rings have appeared sporadically. Although these have been incorporated in descriptions of individual cases, no generalized approach has been proposed (that is, one that would be applicable to all rings, not just the common types). In displaying aortic arch anomalies, previous investigators have emphasized the use of a single frontal cut and multiple parasagittal cuts. Our experience with coronal and axial nuclear magnetic resonance images suggested that a suprasternal sweep, starting in the frontal plane and including (at its most posterior angulation) a plane similar to an axial magnetic resonance cut, would better display the encirclement of the trachea. We review our experience with a standardized tomographic echocardiographic approach from the suprasternal or high parasternal window for the detailed delineation of the superior mediastinal structures.

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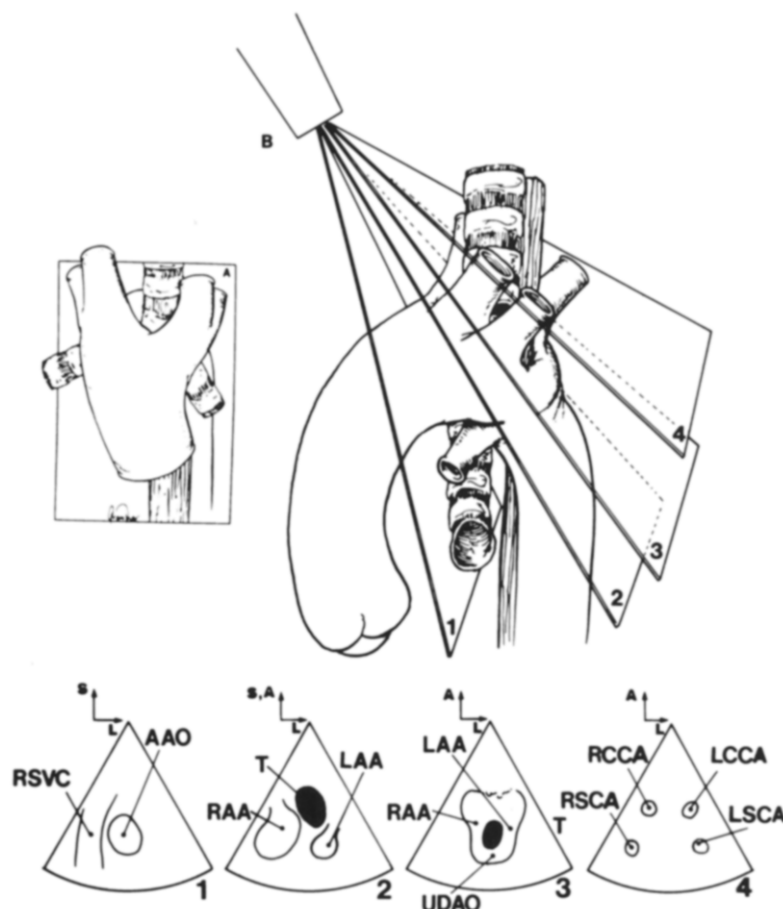
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### Methods

**Study patients.** Between May 1985 and April 1989, 12 patients with a vascular ring (aged 5 to 352 days; median 138) underwent adequate ultrasound examination before surgical repair. Information regarding cardiac catheterization and additional investigative studies was retrospectively reviewed. One of us examined the ultrasound video recordings. The operative anatomy and surgical procedure performed in each case were also reviewed.

**Imaging technique.** A Hewlett-Packard Sonos 500 phased array ultrasound system was used together with 5 MHz short- and medium-focus transducers; Doppler color flow imaging was used in seven patients examined since January 1987. Since June 1988, we additionally used a Hewlett-Packard Sonos 100 mechanical scanner with 7.5 and 5.0 MHz short-focus transducers. Infants were sedated with oral (or rectal) chloral hydrate (60 to 125 mg/kg body weight) before the echocardiographic examination. Although many infants had tachypnea and stridor before referral to the noninvasive imaging laboratory, respiratory depression as an adverse response to sedation was not a problem in this patient group. Patients first underwent assessment of the intracardiac structures by standardized subcostal sweeps, as well as apical and parasternal views.

**Arch structures.** After intracardiac assessment was completed, the patient's shoulders were elevated so that the neck was extended to approximately 30°. Figure 1 depicts the transducer positions and structures intersected by the sectors during the sweep, with the corresponding echocardiographic



**Figure 1.** Line drawing of a simple double aortic arch seen from an anterior perspective (inset A), as well as from the lateral view. Four sectors from the suprasternal sweep (B) have been drawn in, with their corresponding contents in the panels numbered 1 to 4. These correspond to the photographs shown in Figure 2. Because one of the planes is a frontal (coronal) view, we termed this sweep a "frontal" sweep. A = anterior; AA = aortic arch; AAO = ascending aorta; CCA = common carotid artery; L = left; R = right; S = superior; SCA = subclavian artery; SVC = superior vena cava; T = trachea; UDAO = upper descending aorta.

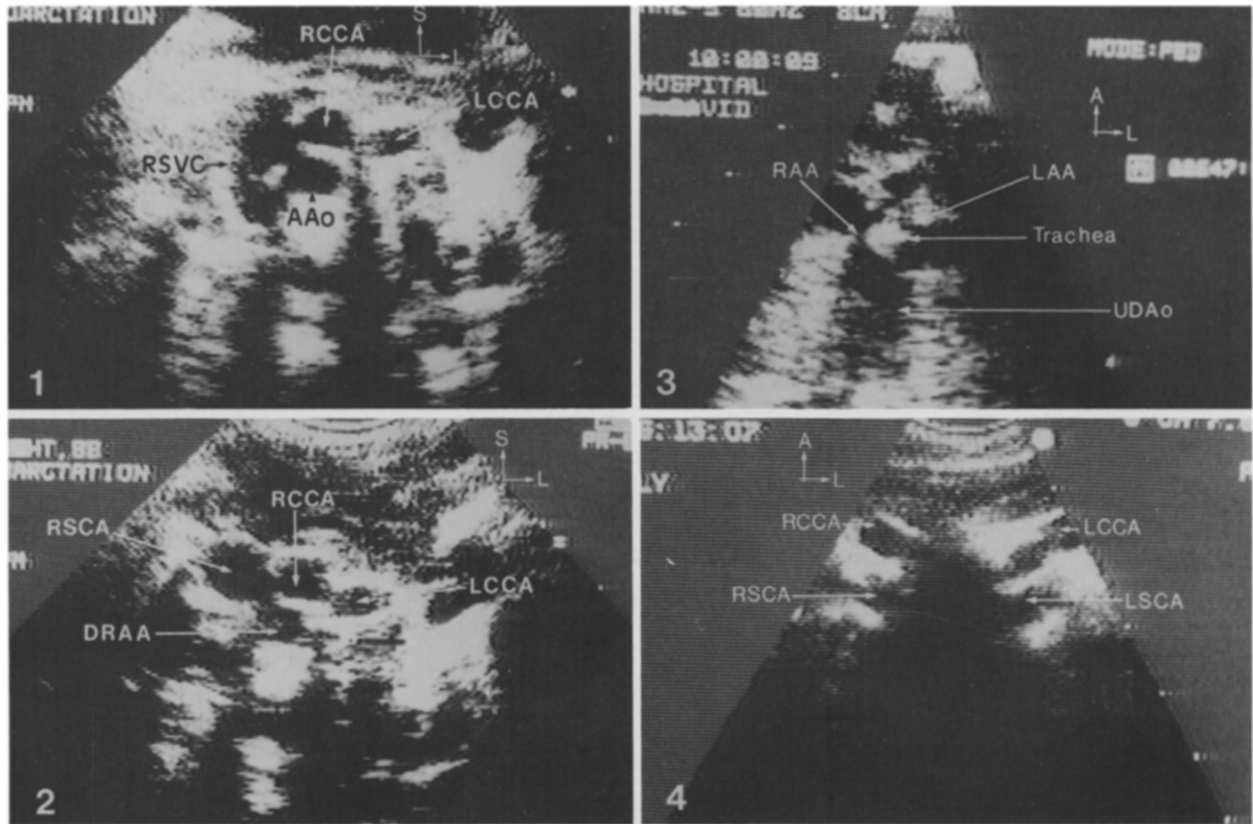
graphic views shown in Figure 2 (as in the case of a typical double aortic arch).

The transducer was positioned in the suprasternal notch or a high parasternal window, and a frontal "reference" view showing the right superior vena cava or bilateral venae cavae was obtained. The transducer plane was then angled anteriorly until the arterial outflow tracts were visualized. From this point, the transducer was slowly swept posteriorly, so that the ascending aorta was displayed in the central region of the sector. This sweep was continued until the first branch of the ascending aorta was visualized.

Particular attention was paid to the lateralization of this vessel in order to assign arch sidedness. If the first vessel coursed to the right, the arch was identified as left-sided, and vice versa. Because airways are usually difficult to identify with ultrasonography, the tracheobronchial structures, which are customarily used as radiographic landmarks to assign arch sidedness, cannot be used routinely in ultrasound imaging. The first branch was followed superiorly and laterally in search of a point where the vessel bifurcated into two vessels of equal or nearly equal caliber. Only if such a bifurcation was demonstrable was the vessel considered to be an innominate artery. Although an innominate artery is typically larger at its origin than the other brachiocephalic

vessels arising from an arch, the size of a vessel was not considered sufficient to designate it as an innominate artery. In the case of either left aortic arch and normal branching or right aortic arch with mirror image branching, a bifurcation point can be found; however, in cases of left aortic arch and aberrant right subclavian artery or right aortic arch and aberrant left subclavian artery, such a bifurcation does not exist because the aberrant subclavian artery arises from the upper descending aorta. Double aortic arch (with or without atresia of one arch) also will not have a bifurcation point in the first arch vessel because each arch gives off a common carotid artery and a subclavian artery. Finally, cases of "isolation of a subclavian artery" will not have a bifurcation point.

The sweep was then continued until a second arch vessel was identified. Its sidedness and course were noted. The third arch vessel and the upper descending aorta were then displayed as the sweep was completed posteriorly. It was important to verify that the third branch coursed laterally instead of directly cephalad to distinguish the subclavian artery from the case in which the third vessel was actually a vertebral artery arising directly from the arch. We also attempted to identify aberrant subclavian arteries and persistent dorsal aortae.



*Sidedness of the upper descending aorta* was noted with the transducer oriented in the frontal plane, centered in the suprasternal notch and directed just anterior to the most posterior angulation of the sweep. In this position, the proximal portion of the upper descending aorta was visualized as a continuation of the distal aortic arch just imaged. If the descending aorta continued on the same side as the distal arch, that portion of the upper descending aorta appeared as a short structure oriented in a cephalocaudal direction. If, however, the upper descending aorta descended on the side opposite to the distal aortic arch, it was visualized as a more elongated structure oriented in an oblique direction from the side of the distal aortic arch to the side of the upper descending portion (Fig. 3).

With the information gained from this sweep, attempts were then made to display arch morphology in other planes. The transducer was placed over the apex of the arch (in a left aortic arch, this was either the suprasternal notch or the high right parasternal window). It was then rotated counterclockwise and angled toward the left hip to achieve a long-axis oblique view, sometimes yielding a "candy cane" appearance. In right aortic arch, the apex of the arch was usually in the suprasternal notch region, and a parasagittal plane was used, with the transducer angled slightly toward the right hip. These planes facilitated complete interrogation of the arch and upper descending aorta with color, pulsed wave

Figure 2. Panels 1 to 4, Successive views from the suprasternal frontal sweep, starting at the most anterior view in the upper left panel, with progressive posterior angulation of the transducer in the subsequent views. These correspond to the sectors drawn in Figure 1. DRAA = distal right aortic arch; other abbreviations as in Figure 1.

Figure 3. Two-dimensional echocardiogram from the frontal sweep at the level of the distal right aortic arch. The arch is crossing the midline to become a left upper descending aorta. The distal arch is seen as an obliquely oriented structure coursing from the upper right toward the lower left of the sector. Abbreviations as in Figure 1.



Table 1. Clinical Profile of 12 Patients

Pt. No.	Age (days)	Operative Anatomy	Echo Diagnosis	Cath	Surgical Approach
1	138	LPA arose from RPA	Complete	No	Sternotomy
2	293	DAA: R > L	Complete	Yes	L thoracotomy
3	86	DAA: R > L, L UDAo	Complete	Yes	L thoracotomy
4	50	DAA: mild narrowing between L SCA and DAo	Complete	No	L thoracotomy
5	129	DAA: LAA atretic between L CCA and L SCA, L UDAo	Incomplete	Yes	L thoracotomy
6	102	RAA: mirror image branching, PDA from L UDAo	Complete	Yes	L thoracotomy
7	247	RAA: anomalous L SCA, LA from L UDAo	Incomplete	No	Sternotomy
8	10	DAA: L > R, LAA atretic between L CCA and L SCA, coarctation distal to R SCA, L PDA	Incomplete	Yes	Sternotomy
9	199	RAA: L UDAo, L LA	Incomplete	Yes	L thoracotomy
10	167	DAA: R > L	Complete	No	L thoracotomy
11	5	DAA: aortic atresia, cervical RAA, LAA atretic between AAO and L CCA, L PDA	Incomplete	Yes	Sternotomy
12	352	DAA: R > L	Complete	No	L thoracotomy

AAo = ascending aorta; Cath = cardiac catheterization; CCA = common carotid artery; DAA = double aortic arch; DAo = descending aorta; Echo = echocardiographic; L = left; LA = ligamentum arteriosum; LAA = left aortic arch; LPA = left pulmonary artery; PDA = patent ductus arteriosus; Pt. = patient; R = right; RAA = right aortic arch; RPA = right pulmonary artery; SCA = subclavian artery; UDAo = upper descending aorta.

and, if necessary, image-directed continuous wave Doppler ultrasound.

**Central pulmonary arteries.** Imaging of the central branch pulmonary arteries and ductus arteriosi was performed starting from the suprasternal frontal view (5). The right pulmonary artery was clearly visualized to just beyond the upper lobe branch by slight angulation of the transducer toward the right chest. By slight counterclockwise rotation into a left oblique view, the left pulmonary artery could be imaged in the left chest adjacent to a left-sided upper descending aorta. In a patient with a right upper descending aorta, the right pulmonary artery occasionally lies adjacent to it.

*In the case of pulmonary vascular sling*, the origin of the left pulmonary artery was displaced to the right, and appeared to arise as a posterior branch of the right pulmonary artery. It then curved toward the left chest, passing around the distal trachea and proximal right mainstem bronchus. In this situation, the left atrial appendage, located anterior to the distal trachea and adjacent to the main pulmonary artery, may be confused with a normal left pulmonary artery; however, the coincident presence of an abnormal origin of the left pulmonary artery from the posterior right pulmonary artery and its retrotracheal course are pathognomonic for a pulmonary vascular sling.

**Ductus arteriosi.** The suprasternal frontal sweep just described and high parasternal parasagittal views (11) were utilized, keeping in mind the potential locations of the patent

ductus arteriosus accompanying each aortic arch configuration. The typical positions for ductus arteriosi in a patient with a left aortic arch are the upper descending aorta (left ductus) and innominate artery (right ductus); an atypical course for a right ductus in a left aortic arch runs from the left upper descending aorta to the right pulmonary artery by way of a persistent right dorsal aorta.

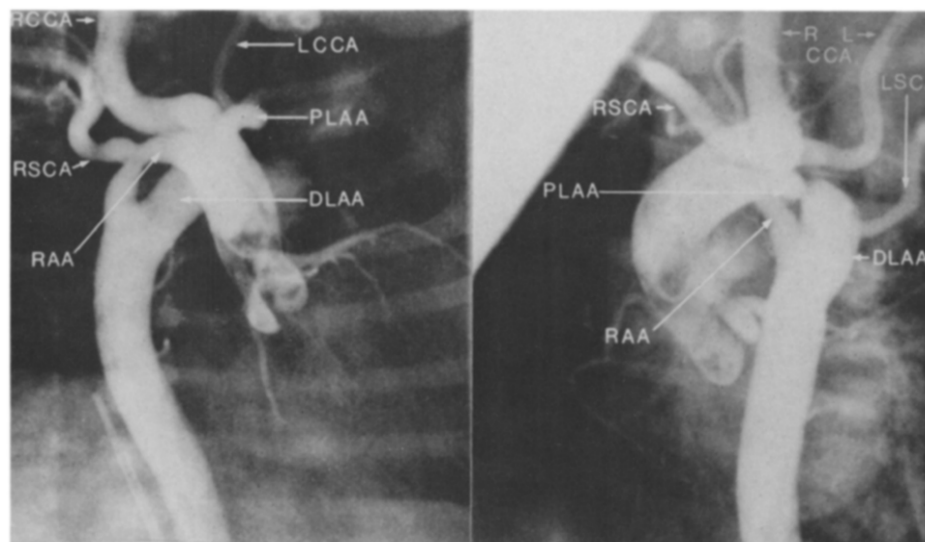
In left aortic arch with an aberrant right subclavian artery, the typical positions for ductus arteriosi are the upper descending aorta (left ductus) and the right dorsal aorta component of the proximal right subclavian artery (right ductus). The most common positions for ductus arteriosi in right arch with mirror image branching and right arch with aberrant left subclavian artery can be extrapolated from the preceding discussion of left arch variants. In double aortic arch, the ductus arteriosus sites are the left upper descending aorta (left ductus) and right upper descending aorta (right ductus).

## Results

Table 1 summarizes the findings for each of the patients. Nine patients had a barium esophagogram performed; each was suggestive for the presence of a vascular ring. Seven patients underwent cardiac catheterization.

**Ultrasonography.** Two-dimensional ultrasonography alone successfully delineated all of the components of the vascular rings disclosed at surgery in 7 of the 12 patients. Of

**Figure 4. Patient 8. Aortograms.** Left panel, Right anterior oblique view of an aortic root injection. Right panel, Left anterior oblique view of the same injection. There is atresia of the left aortic arch between the left common carotid artery and left subclavian artery origins. DLAA = distal left aortic arch; PLAA = proximal left aortic arch; other abbreviations as in Figure 1.



the five patients with an incomplete diagnosis, two had right aortic arch with left upper descending aorta, which was correctly identified by ultrasonography. In one of these patients, no ductus arteriosus could be identified; however, a left ligamentum arteriosum was discovered intraoperatively. In the other, an anomalous left subclavian artery was detected on echocardiography; however, an accompanying left ligamentum arteriosum was not recognized. The other three patients with an incomplete diagnosis had more complex arch anatomy. One patient with double aortic arch proved to have atresia of the left aortic arch between the left common carotid artery and left subclavian artery; ultrasonography had suggested narrowing, but not atresia. In another, the anatomy consisted of aortic atresia, double aortic arch with cervical right arch, atresia of the left arch proximal to the left common carotid artery, and a left ductus arteriosus. The cervical extent of the right aortic arch was not appreciated by ultrasound. The fifth patient had double aortic arch with hypoplasia of the right aortic arch (and discrete coarctation just distal to the origin of the right subclavian artery). An atretic segment of the left aortic arch between the left common carotid artery and the left subclavian artery had not been displayed by ultrasonography; the latter yielded the diagnosis of right aortic arch with aberrant left subclavian artery and left ductus arteriosus.

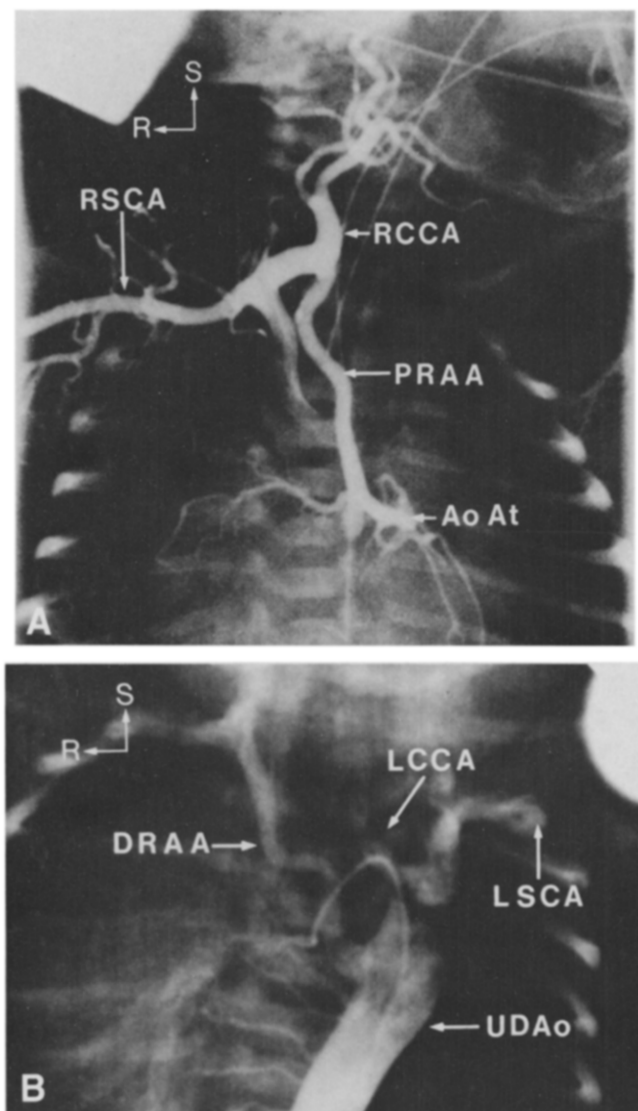
*In four of the five patients with an incomplete diagnosis, the area not displayed had an atretic lumen.* Only the patient with a cervical arch had an incomplete study in which a patent vascular segment was not identified.

**Angiography.** Five of the seven patients who underwent cardiac catheterization had double aortic arch. Two of these had simple aortic arch with dominance of the right arch in one and almost equal caliber in the other; however, the other three lesions were complex. One patient had atresia of the left aortic arch between the left common carotid artery and

the left subclavian artery; another had hypoplasia of the right aortic arch (with a discrete coarctation just distal to the origin of the right subclavian artery) and atresia of the left aortic arch between the left common carotid artery and the left subclavian artery with left patent ductus arteriosus (Fig. 4). The third patient had aortic atresia with a cervical right aortic arch, atresia of the left aortic arch proximal to the left common carotid artery, and a left patent ductus arteriosus (Fig. 5).

*The other two patients who underwent cardiac catheterization had right aortic arch with a left upper descending aorta.* One had mirror image branching and a left patent ductus arteriosus, and the other had a left ligamentum arteriosum arising from the persistent left dorsal aorta.

**Additional imaging.** The addition of a second noninvasive technique allowed sufficient information to be obtained for accurate description of the anatomic features of the vascular ring. This second noninvasive procedure included a suggestive barium esophagogram; however, two of the more recent patients (Patients 7 and 9) also had spin echo nuclear magnetic resonance imaging performed. Each of these patients had a right aortic arch. Patient 7 had an anomalous left subclavian artery (Fig. 6, right lower panel), with a left ligamentum arteriosum noted at surgery. Patient 9 had a left descending aorta and a left ligamentum arteriosum arising from the left upper descending aorta (Fig. 6, left lower panel). In both patients, the presence of a vascular ring had been suggested by barium esophagogram (Fig. 6, upper panels); however, ultrasonography had been unable to define the structure connecting the anterior and posterior segments of the ring. Nuclear magnetic resonance imaging could not demonstrate the ligamentum arteriosum in either patient; however, the presence of a diverticulum of Kommerell in one patient and an aortic ductus dimple in the other suggested the presence of a left ligamentum arteriosum.



**Figure 5.** Patient 11. Aortograms. **A**, Frontal view taken from an upper descending aorta injection. This frame demonstrates several of the anatomic features that were peculiar to this patient, including aortic atresia (Ao AT) and hypoplasia of the cervical right aortic arch. **B**, Frontal view taken from an upper descending aorta injection. This frame demonstrates the distal right cervical aortic arch (DRAA), as well as the left common carotid and subclavian arteries arising from the distal left aortic arch. PRAA = proximal right aortic arch; other abbreviations as in Figure 1.

## Discussion

### *Previous Studies on Diagnoses of Vascular Ring and Aortic Arch Anatomy*

As recently as 1980, Ergin et al. (12), in a case description of left aortic arch with right upper descending aorta and right ligamentum arteriosum, recommended that "barium swallow, magnification airway roentgenograms and diagnostic

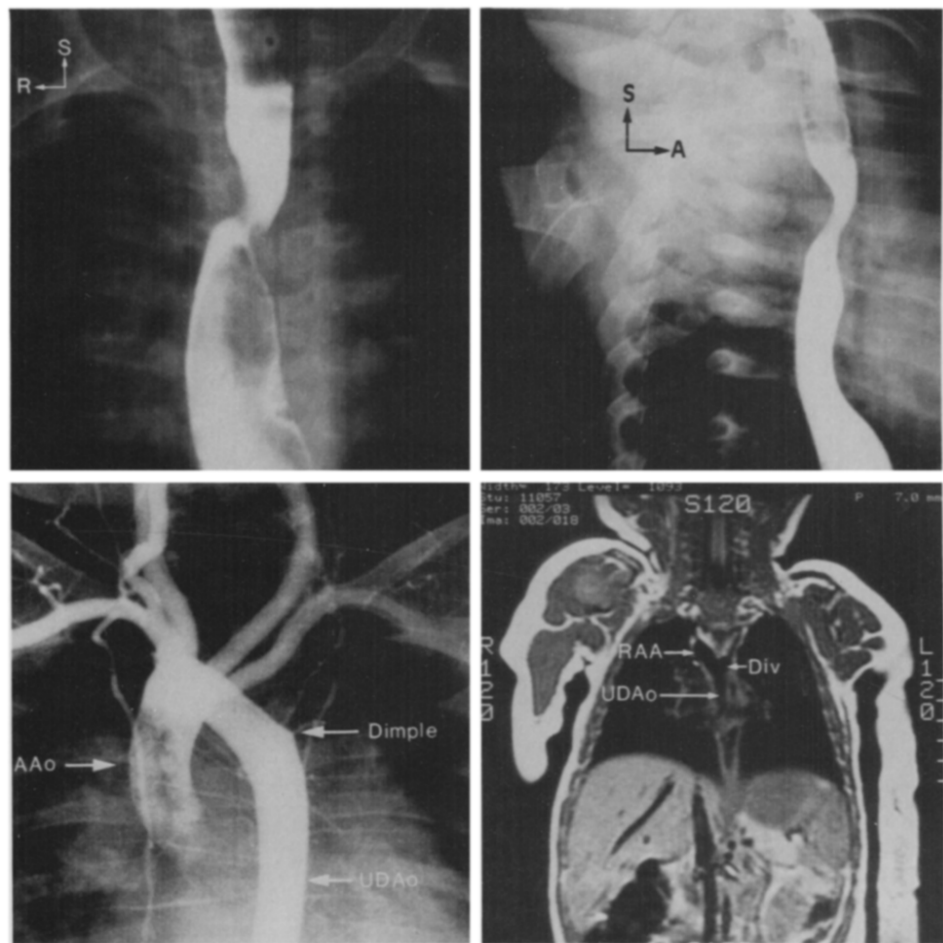
cardiac catheterization, including aortography, should be routine in the preoperative workup" of any patient suspected of having a vascular ring. In 1981, Snider and Silverman (5) introduced suprasternal "long-axis" and "short-axis" echocardiographic views for the imaging of extracardiac structures. In a case series description in 1982, Sahn et al. (1) combined subcostal coronal echocardiographic imaging with intravenous digital subtraction angiography in the display of double aortic arch. Variations in transducer positioning (suprasternal or parasternal) and orientation were subsequently reported (2,6), providing the echocardiographer with a number of single-plane approaches to imaging of the aortic arch, whether left or right. Unfortunately, these approaches depend on the subjective impression of a deviation in arch position from normal; moreover, because no fixed landmarks are used, an observer reviewing a videotaped study cannot easily deduce the exact planes used by the ultrasonographer.

**Determining arch sidedness.** Appreciation of the relative position of the aorta to the echogenic air-filled trachea in right and left aortic arches led to a description of a parasternal transverse view for determining arch sidedness (7). Similarly, Huhta et al. (8) ascribed arch sidedness by direct visualization of the esophagus during the swallowing of liquid in sagittal scans, and subsequent location of the transverse aorta by scanning to the right and left of the esophagus. As an alternative, they proposed the use of the thoracic vertebral bodies as midline landmarks. Huhta (9) later described the administration of a small amount of a carbonated liquid, which allowed easy and rapid recognition of the esophagus. He also noted that the innominate artery arises from the side opposite the arch; its display may be helpful in the assignment of arch sidedness. However, he stated that in the case of anomalous origin of the subclavian artery, this method is not confirmatory of the side of the arch. Kveselis et al. (10) also noted that the diagnosis of a right aortic arch could be suspected when the first branch coursed leftward and superiorly.

Smallhorn et al. (11) reported on the suprasternal (or high parasternal) parasagittal view for the assessment of a left-sided patent ductus arteriosus in a patient with a left aortic arch. Enderlein et al. (3) described the use of suprasternal imaging for the description of double aortic arch anatomy utilizing a standard "long-axis" view ("candy cane" configuration) to visualize the left arch, with rotation of the transducer to locate the right arch. Finally, in a single case report (4), the addition of color flow mapping further allowed the operator to document the pattern of circulation in the vessels of the double arch. Although noncontiguous planar techniques may be useful for the specific cases described and applicable to patients with relatively uncomplicated anatomy, a generalized approach using a sweep comprising contiguous planes offers the most understandable, reproducible display of aortic arch anatomy in complex cases.



**Figure 6.** Upper panels, Patient 9. Frontal (left) and right (right) oblique views of the barium esophagogram. Left lower panel, Patient 9. Frontal projection from the aortic root angiographic injection. This clearly demonstrates the presence of a right aortic arch with mirror image branching, retroesophageal course of the distal arch and left upper descending aorta. The aortic end of the ligamentum arteriosum produces a dimple at the proximal end of the left upper descending aorta. Right lower panel, Patient 7. Frontal view from the nuclear magnetic resonance image at the level of the distal right aortic arch. This demonstrates the distal end of the right aortic arch and left upper descending aorta, the diverticulum (Div) of Kommerell and the origin of the anomalous left subclavian artery. Abbreviations as in Figure 1.



The use of standard subcostal and parasternal short-axis views was described by Yeager et al. (13) for the imaging of pulmonary vascular sling. Although the suprasternal window was not used in their case series, these authors speculated that its use may facilitate display of the anomalous branching of the left pulmonary artery.

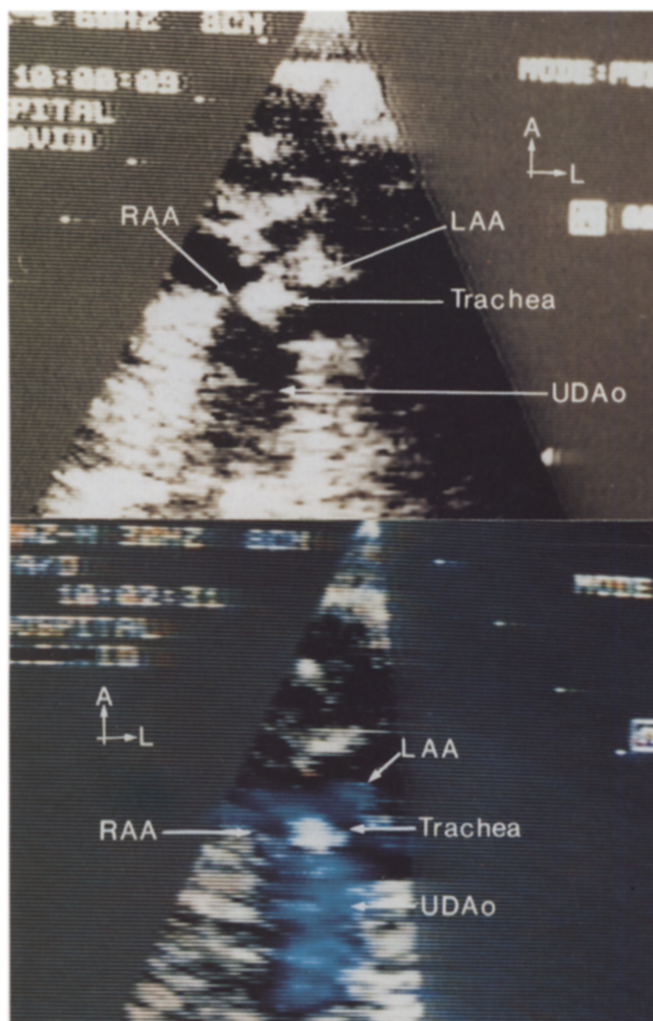
### *The Present Imaging Method*

Using the technique described, we were able to reliably document the anatomy and spatial relations of the vessels in this group of patients with a vascular ring. Although more time-consuming than using single-plane views, the use of multiple contiguous cross-sectional views in a continuous sweep gives the examiner (and others later reviewing a video recording) a mental three-dimensional image of the mediastinal vascular structures. The addition of Doppler color flow information was very helpful in tracing multiple vascular structures involved because the vascular courses were tortuous and the near-field (0 to 3 cm depth) resolution with phased array instrumentation is low (even using a short-focus 5 MHz transducer). With posterior angulation, the suprasternal frontal sweep provided visualization of the

encirclement of the air-filled trachea, which was also emphasized further when Doppler color flow imaging was added (Fig. 7). In patients with double aortic arch, the arch vessel origins form the corners of a rectangle (Fig. 2, lower right panel).

**Sedation.** Ultrasonographic assessment of vascular rings is dependent on the adequate imaging of structures and is impossible without patient compliance throughout the examination. Adequate sedation is imperative. Although patients with a vascular ring are frequently referred because of signs of upper airway compression, respiratory distress has not occurred using sedation in these doses. Patients who are unresponsive to chloral hydrate can be given pentobarbital (3 mg/kg orally) or midazolam (0.1 to 0.2 mg/kg intravenously).

**Surgical approach.** Detailed description of the anatomy of the arch (or arches) and branches is necessary for a rational surgical approach. With superb imaging, decisions regarding surgical repair can be made without the need for more invasive tools, including angiography. We agree with previous investigators (14,15) who pointed out that the surgical axiom that all vascular rings can be divided through



**Figure 7.** Patient 10. Upper panel, Two-dimensional echocardiogram from the suprasternal frontal sweep. The transducer has been angled posteriorly to image the encirclement of the trachea (the densely echogenic structure) and esophagus by the right and left segments of the double aortic arch, as well as the upper descending aorta. Lower panel, A similar view with color Doppler information added. This more dramatically outlines the course taken by each segment of the double aortic arch. LAA = left aortic arch; RAA = right aortic arch; other abbreviations as in Figure 1.

the left side of the chest is false. Surgical repair in four of our patients was performed through a median sternotomy.

**Limitations.** Although ultrasonography adds considerable detail to the anatomic information that can be gathered by barium esophagography, there are limitations. Because it displays structures with fluid-filled lumens, it does not delineate nonvascular structures such as ligamentum arteriosum or a fibrous cord linking the anterior and posterior segments of an atretic arch. It is difficult to distinguish between atresia of one segment of a double aortic arch and a complete interruption at the site. In the adjacent arch segments, there may, however, be clues to the presence of

these ligamentous structures, such as a diverticulum of Kommerell, persistent dorsal aorta or an aortic dimple (from tethering by the fibrous attachment). In two of our patients with ligamentum arteriosum, other imaging techniques (angiography and nuclear magnetic resonance imaging) were able to display these clues, allowing us to infer the presence of ligamentous structures. Many patients will be referred to the cardiologist after initial assessment of signs or symptoms of airway compression, often having had suggestive barium esophagography. In a symptomatic patient, it is sufficient to display the three luminal segments of a vascular ring by ultrasonography when this is combined with a technique, such as barium esophagography, that provides information about the adjacent nonvascular structures. Cervical arches present a special challenge to the ultrasonographer; further experience should provide a reliable approach.

*In patients whose body characteristics preclude adequate ultrasound imaging, computed tomography (16) and nuclear magnetic resonance imaging (17) are particularly useful for detailed anatomic delineation. These techniques offer the advantage of displaying vascular, airway and esophageal structures simultaneously in a wide field of view. The creation of three-dimensional reconstructions from nuclear magnetic resonance images has proved to be a very powerful imaging tool.*

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